Percutaneous Ventricular Septal Defect Closure

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Abstract

Ventricular Septal Defects (VSD) are common in early childhood but the prevalence in adulthood is low due to spontaneous closure. Echocardiography can assess the location and size of the VSD, the direction of the intra-cardiac shunt, the effect of the VSD on the cardiac chambers and other associated anomalies. Percutaneous closure of the VSD has become an alternative to surgery. The location of the VSD determines it's feasibility for device closure. Echocardiography guidance is critical for the safe and successful VSD closure as well for the follow up evaluations.

Keywords

Ventricular Septal Defect (VSD) • Percutaneous closure • Echocardiographic guidance • Kirklin VSD classification

A ventricular septal defect (VSD) is an abnormal communication in the ventricular septum between the left and right ventricles. VSDs can be either isolated or occur in combination with other cardiac defects. They are common congenital heart anomalies in early childhood, occurring in about 3–3.5 per 1,000 live born infants. In the adult population, due to the spontaneous closure of small defects, the prevalence is lower, occurring in about 0.3 per 1,000 persons. According to the location of the defect in the ventricular septum, VSDs can be classified as either membranous or muscular defects [1, 2]. They also can be categorized in terms of their size in relation to the aortic annular diameter. Small VSDs are those that measure ≤25 % of the aortic annular diameter, moderate ones are those that measure between 25 % and 75 % of the aortic annular diameter, and large VSDs measure ≥75 % of the aortic annular diameter. Small defects are usually restrictive, with a Qp:Qs ratio <1.4. In these cases, the pulmonary artery, left ventricle, and left atrium typically remain normal sized. Moderate defects can cause mild-to-moderate left ventricular, left atrial, and pulmonary artery enlargement with a Qp:Qs ratio ranging from 1.4 to 2.2. Large nonrestrictive VSDs with Qp:Qs ratios 2.2 can cause volume overload and dilation of the left-sided cavities and pulmonary artery. Many of these patients will develop pulmonary vascular disease leading to a reduction in left-to-right shunting or possible shunt reversal due to the occurrence of Eisenmenger physiology [3].

The initial echocardiographic evaluation of a patient with suspected or known VSD should include assessment of the location and size of the VSD, the direction and magnitude of intracardiac shunting, the size and function of the cardiac chambers, right and left ventricular outflow measurements, valvular morphology and function (including assessment for aortic valve prolapse), pulmonary artery pressures, and the presence of any other associated cardiac anomalies [4].

One of the most important observations is the anatomic location of the defect, as it defines the feasibility of device closure. The Kirklin classification describes the location of the VSD in relation to the ventricular septum when the defect is viewed from the right ventricle. A type I defect refers to a subarterial defect, which is located at the anterosuperior part of the ventricular septum; type I defects are seen in about

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5 % of cases in the United States. A type II defect refers to a perimembranous VSD that is located in the middle of the upper portion of the ventricular septum in close proximity to the aortic valve. Type II defects are present in 75 % of patients with a VSD; these are associated with a high incidence of spontaneous closure. A type III defect is a perimembranous VSD with inlet extension; it is present in the posterior portion of the ventricular septum in close proximity to the tricuspid valve, and it is present in about 5 % of patients. A type IV defect is a muscular VSD. It is present in 10–15 % of patients and is most commonly present at the apical septum. Muscular VSDs can be single or multiple; in the latter case they may be referred to as a "Swiss cheese" VSD due to their appearance.

First introduced by Lock and colleagues, transcatheter VSD closure has become an alternate approach to surgery [5]; early efforts involved the use of a Rashkind double-umbrella device. The use of Amplatzer devices designed for muscular and perimembranous VSDs have been associated with a 100 % success rate in one study with up to 12 months of post-procedural follow-up [4]. Important complications of device closure include device embolization, hypotension, bleeding, and conduction abnormalities. The reported rate for muscular VSD closure complications is 10.7 %. Perimembranous VSD closure is associated with even higher rates of complications, with considerable risk of damage to the aortic valve, tricuspid valve, and conduction system. Because of the high risk of complications, perimembranous VSD closure has been partially abandoned [6, 7]. In general, device closure carries a class IIb indication (level of evidence C). Device closure can be considered as a treatment option if the VSD is not close to the tricuspid and aortic valves and if it is associated with severe left-sided chamber enlargement or pulmonary hypertension [8].

Although transcatheter closure is an established method of treating selected congenital VSDs, there is less clinical experience regarding transcatheter closure of post-infarction VSDs. The procedure is considered difficult because the margins of the defect may have necrotic borders upon which the device may not anchor and the patients tend to be critically ill at the time of the procedure [9]. Even in successful cases, the mortality remains high due to advanced patient age, comorbidities, severity of coronary artery disease, hemodynamic instability, and procedural complications. In a recent clinical trial, the 30-day survival was 35 % and the long-term survival was 30 % [10, 11].

Percutaneous closure is typically performed under general anesthesia and echocardiographic guidance. Either a transthoracic (TTE) or transesophageal (TEE) approach can be used. Echocardiography will guide the proceduralist on the number and size defects, their location, and their position with respect to the valves and other nearby structures. In some cases the right internal jugular vein, femoral vein, and femoral artery are all accessed for the procedure. Right internal jugular access is especially helpful in patients with a mid, posterior, or apical VSD. For mid-muscular, posterior, or apical VSDs, a left ventricular angiogram is performed in the hepatoclavicular view (35° left anterior oblique [LAO]/35° cranial) to delineate the location, size, and number of VSDs; for anterior defects, the long axial oblique (60° LAO/15° cranial) view is used. Either a Judkins right coronary artery catheter or a Cobra catheter can be used to cross the VSD from the left ventricular side. Typically a right heart catheterization is also performed prior to crossing the VSD; once the VSD is crossed from the left ventricular side, a long guidewire can be used to traverse the lesion and a snare device in the right heart can be used to pull back the long guidewire, externalizing it with respect to the venous access site. This in effect creates an "arteriovenous (AV) loop" that serves as a stable rail from the venous circulation to the arterial circulation across which the device can be deployed. As the device is passed from the venous circulation into the right ventricle and across the VSD, both echocardiography and angiography can be used to verify device position, as well as the deployment of the device discs, the release and stability of the device, and the presence of any residual shunting.

The distance from the aortic valve leaflets is an important measurement that should be determined before the procedure, as the absence of tissue separating the aortic valve leaflets from the orifice can result in complications. In general, the defect should be at least 2 mm away from the aortic valve. Additionally the presence of accessory tissue around the defect must be verified, as the prosthesis must be attached to the muscular septum. Some defects have large amounts of surrounding accessory tissue, forming an aneurysm, which significantly reduces the size of the original orifice. These defects may be occluded with smaller devices, which are positioned inside the aneurysm sac and do not compromise tricuspid or aortic valve function. The presence of aortic valve prolapse, especially of the right coronary leaflet, can be seen in 2–7 % of VSDs, leading to aortic insufficiency. Although this may be a relative contraindication for a percutaneous procedure, there are cases of device implantation performed under these circumstances that have been carried out safely and effectively. Echocardiography should also define the presence of any associated defects, such as a subaortic membrane or a right ventricular muscle band; these may require a surgical approach. Lesions such as pulmonary or aortic valve stenosis, persistent ductus arteriosus, and aortic coarctation can also be treated percutaneously, depending on their anatomical features [12–14].

Muscular VSDs located in the trabecular and apical regions of the septum can also be treated percutaneously. Although there are fewer anatomical variations, the

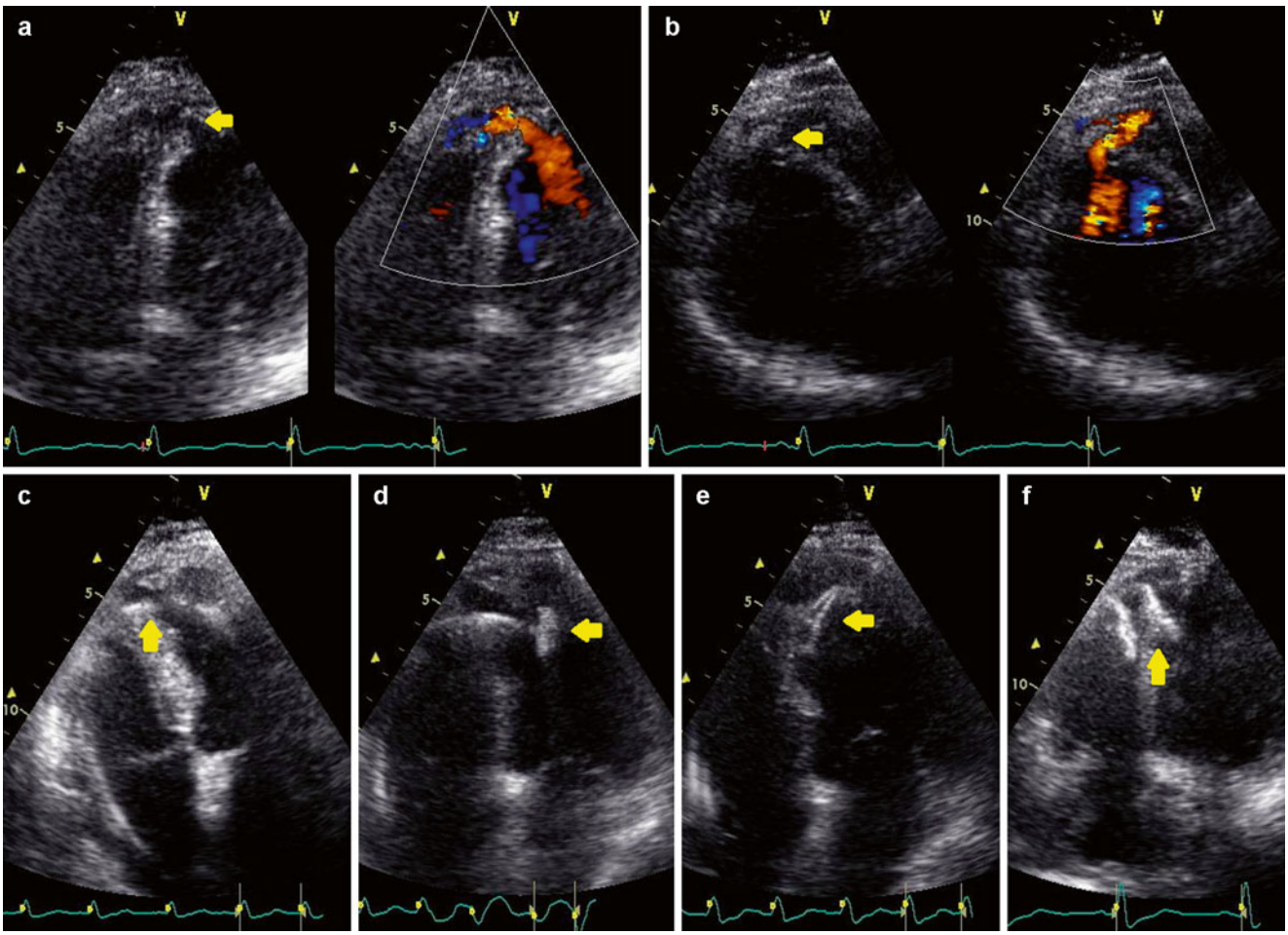

Fig. 10.1 Transthoracic echocardiographic imaging demonstrating percutaneous closure of a post-infarct apical muscular VSD. Panels (**a**, **b**) were obtained prior to the procedure, demonstrating the presence of VSD from apical four chamber (panel **a**) and off-axis parasternal long axis (panel **b**) views (*arrows*). The corresponding color Doppler images demonstrate flow across the VSD. Panel (**c**) demonstrates the course of the AV loop of guidewire (*arrow*), which is created when the long guidewire that had been passed through the VSD from the left ventriclular side is snared from the right ventricular side and externalized. Panel (**d**) demonstrates deployment of the left ventricular disc, which is subsequently pulled back until it abuts the ventricular septum (panel **e**). Finally, panel (**f**) shows deployment of the right ventricular disc and release of the device, which is well seated against the apical septum

presence of multiple orifices is common, particularly in the ventricular apex. When there is a single orifice, the Amplatzer device is the best choice, as it has small retention discs that occupy a small amount of space on the interventricular septum [15].

Recently, a hybrid approach for the management of very young patients (<6 months of age) with large muscular VSDs has been utilized. These patients are taken to the operating room for a thoracotomy to expose the heart. Through a right ventricular free wall puncture, a guidewire is introduced into the left ventricle through the VSD. A short sheath is then advanced over the guidewire allowing for device positioning. The procedure is performed under TEE monitoring. This innovative approach, in which the interventionalist, echocardiographer, and cardiac surgeon all work together, prevents the deleterious effects of extracorporeal circulation, and results in occlusion rates >90 %.

Echocardiography During Percutaneous VSD Closure

The procedure can be performed with TTE or TEE guidance. Although echocardiographic imaging is important for guiding the procedure, fluoroscopy and angiography are also utilized during the intervention [16–18]. If TEE is being used, intermediate transducer image planes between 0 and 180° can usually provide visualization of the VSD and device positioning. Similar to ASD closure, echocardiography is used for orifice measurement and device size choice, monitoring the guidewires, catheters, and sheaths as they are positioned inside the heart, and determining the location of the device discs before and after deployment. The echocardigrapher must also carefully evaluate the aortic and tricuspid valves, as well as evaluating the interventricular septum for the detection and grading of possible residual shunts.

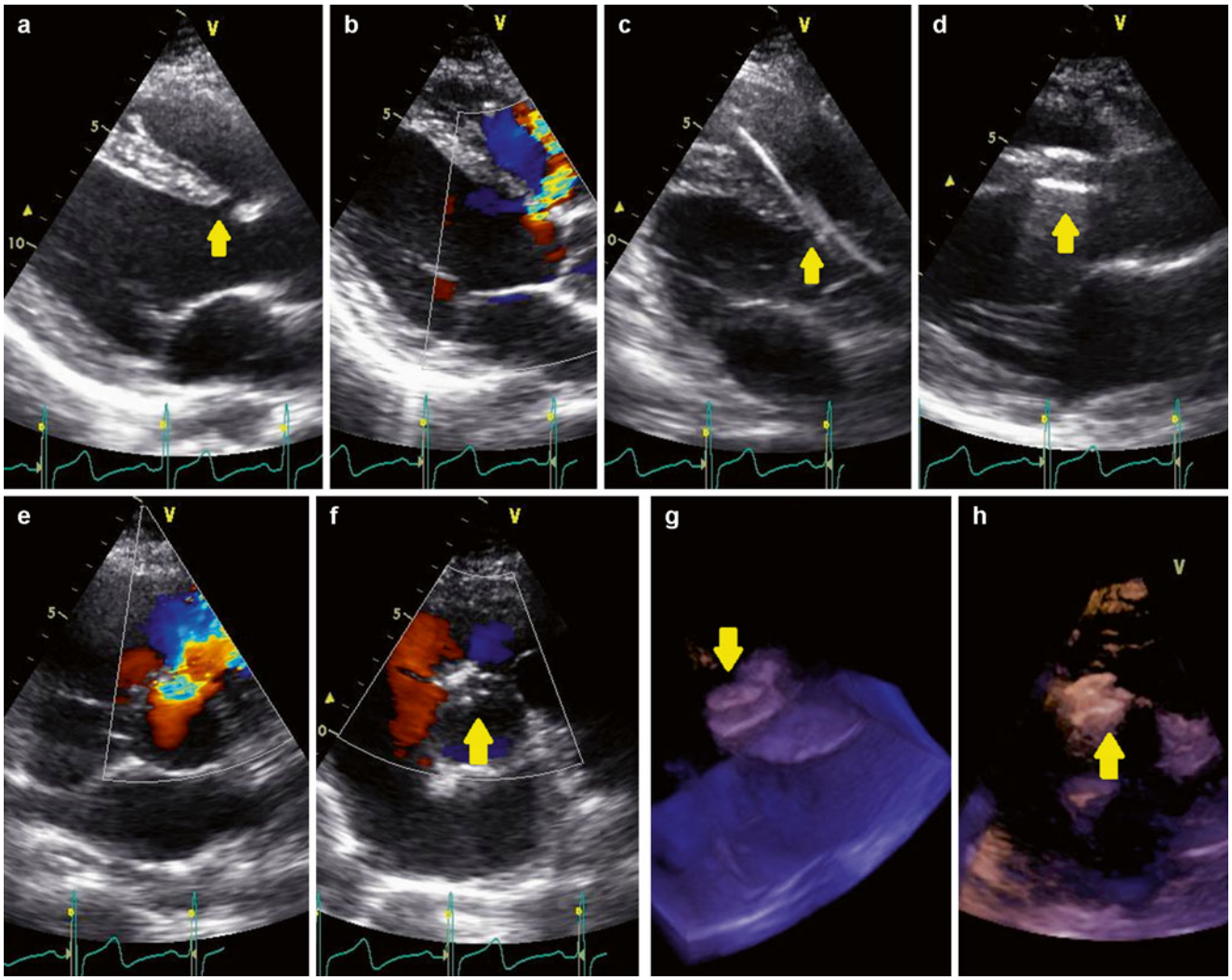

Fig. 10.2 Transthoracic echocardiographic imaging demonstrating percutaneous closure of a congenital perimembranous VSD. Panels (**a**, **b**, **e**) were obtained prior to the procedure, demonstrating the presence of VSD from parasternal long axis (panels **a**, **b**) and short axis (panel **e**) views. The color Doppler images in panels (**b** and **e**) demonstrate left to right flow across the VSD. Panel (**c**) demonstrates the AV loop of wire (*arrow*), passing from the aorta, into the left ventricle, across the VSD, and into the right ventricle. Panel (**d**) demonstrates in the parasternal long axis view the deployed device, which is well seated against the interventricular septum. Minimal residual device flow was visualized (panel **f**), with a substantial decrease noted between panel (**e**) (pre-procedure) and panel (**f**). Panels (**g**, **h**) show three-dimensional images of the deployed device (*arrows*)

Figures 10.1 and 10.2 demonstrate guidance of percutaneous closures of post-infarct and congenital VSDs, respectively, using TTE. Regardless of the approach used, the image quality is dependent on the experience of the echocardiographer as well as the patient's echocardiographic windows. If the patient is intubated, short periods of held ventilation may help stabilize the position of the heart in the chest to allow for better quality images. While standard imaging planes are certainly helpful, the echocardiographer should not be committed only to these and should try alternate planes and/or off-axis imaging as needed to obtain optimum visualization.

Follow-Up Echocardiography After Percutaneous VSD Closure

After percutaneous VSD closure is performed, patients are followed with serial clinical assessments and TTEs. Several parameters must be evaluated, including the left-sided cavity dimensions (which typically return to normal in the first year after closure), the positioning of the device, the presence and severity of any residual shunts, the evaluation and measurement of a possible gradient in the left ventricular outflow tract, and the function of the aortic and tricuspid valves.

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